221 Relationship between growth, nutritional status and pulmonary function in children and adolescents with Cystic Fibrosis

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Background: Nutritional status and lung function are associated with survival in Cystic Fibrosis (CF).
Aim: To determine the relationship among growth, nutritional status and pulmonary function in children and adolescents with CF.
Methods: A cohort of 48 children aged 6 to 10 was followed every 4 months over a 13-year period at Thessaloniki CF center. Weight-for age, (WFA), height-for-age (HFA), percent ideal body weight (%IBW) and pulmonary function test (PFT) results were recorded.
Results: The prevalence of abnormal weight for height (<90%) increased with age from 19% in children aged <7 years to 40% in adults with CF. Patients with malnutrition had significantly lower mean values of Forced Vital Capacity (FVC) and Forced Expiratory Volume in one second (FEV1) ($p < 0.05$). Longitudinal follow-up showed that malnourished patients had significantly worse lung function than their normally nourished counterparts and a greater yearly loss of FEV1.
Conclusions: Our data emphasize the close relationship between nutrition, lung function and clinical course in CF.

222 Lung transplantation in CF patients on mechanical ventilation and ECMO

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Background: It is not recommended to perform lung transplantation (Tx) on CF patients while on ventilator. We have successfully transplanted 3 CF patients on ventilator and extra-corporeal membrane oxygenation (ECMO). Mean follow up 835 days, all alive.
Description: Our patients were between 24 and 44 yo, 1 F. One patient was on waiting list (WL) and two were transplanted under acute circumstances. All were colonized with *Pseudomonas aeruginosa* (PA) and one also *C. acidovorans*.
The female patient had a pneumothorax with accompanying subcutaneous emphysema. Last measured FEV1 was 31% of predicted. She was treated with high frequency oscillating ventilation (HFO) combined with controlled mechanical ventilation (MV) for 22 days and 10 days on ECMO before Tx.
One patient was slowly deteriorating and on WL for 49 days when hospitalised. FEV1 21% of predicted. On non-invasive ventilation for 30 days. On ECMO with PCO_2 17 kPa the day before Tx.
Our last patient was rapidly deteriorating with pneumonia and large hilar nodes. Accepted for Tx after 21 days in hospital. On MV with PCO_2 15 kPa and pH 7.25. Tx after 10 days on MV and 6 days on ECMO.
All patients were treated with airway clearing therapy (ACT) sessions 2–4 times per day combined with inhalations. Bronchoscopy for bronchial clearing was performed as needed, usually once daily. The amount of evacuated sputum varied between 100 mL and 500 mL per session. ACT and bronchoscopy were often done at the same time. The patients were put on ECMO when we could not clear hypercapnia and reach acceptable oxygenation in spite of the treatment.
Conclusion: Tx on CF patients on MV and ECMO is possible with good results. ACT has to be performed regularly with competent physiotherapists. Cooperation between the involved specialties is important.

223 Comparing the clinical evolution of Cystic Fibrosis patients with meconium ileus and with nonmeconium ileus diagnosed under two months

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Meconium ileus (MI) is the earliest symptom of cystic fibrosis (CF) and is the presenting feature in 6–20% of causes of cystic fibrosis.
Aim: To show the difference between the patients with CF with MI and with nonmeconium ileus (nMI) for hospitalization for pulmonary complications and pseudo-Bartter syndrome (PB), colonization with *Pseudomonas aeroginosa* (Pa) and *Staphylococcus aureus* (Sa), the status of growth with Body Mass Index (BMI).
Method: We have performed a retrospective study for the previous data of 195 patients with CF who were treated in Hacettepe University Hospital between January 1987-December 2005. We have identified 14 cases of classical CF with MI by using radiological, sweat and genotype testing and compared them with 23 CF patients with nMI, who were diagnosed under two months old, for hospitalization for pulmonary complications and for PB, for colonization with Pa and Sa, for the status of growth.
Results: Of 14 CF patients with MI one patient had Pa colonization, and of 23 CF patients with nMI two patients had Pa colonization and two patients had Sa colonization ($p > 0.837$). Eight CF patients with MI and 11 CF patients with nMI have been hospitalized with pulmonary infection ($p > 0.916$). Seven CF patients with MI and 13 CF patients with nMI have been hospitalized with PB ($p > 0.102$). We also could not find a difference between two groups for the last BMI results ($p > 0.975$).
Conclusion: We did not find a significant difference between two groups for the respiratory status, BMI, hospitalization for PB, and colonization with Pa and Sa.

224 Adults with Cystic Fibrosis: a single centre experience over five years

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Aim: To better characterise the patients of our adult CF program.
Method: Retrospective analysis of patient charts over the last 5 years.
Result: The actual number of patients in our program is 24 (11 women and 13 men). Median age is 25.8 y (range 18.8–50.6). Most patients have moderate to severe obstructive lung disease with a median FEV1 of 1.68 litre (0.67–4.13), 54.5% predicted (19–100%). Actual body mass index is median 20.8 kg/m^2 (17.0–28.2). There was no significant change in BMI over the last year. Fourteen (58.3%) have chronic infection with *P. aeruginosa* (PA), 6 (25%) have no infection with PA, and 4 (16.7%) grow PA intermittently. No patient had an infection with *B. cepacia*. Three (12.5%) patients had an infection with an atypical mycobacterium. Five (20.8%) patients have CF-related diabetes mellitus and 2 have significant osteoporosis. Intermittent haemoptoe was present in 6 (25%), pneumothorax in 2, and classic allergic bronchopulmonary aspergillosis in 2. Since the start of the adult CF program, no patient died; one had successful lung transplantation. However, lung function decreased significantly over the last year: loss of FEV1 ($p < 0.01$): median –125 ml (range –700 to +170 ml). This was independent of gender, the presence of PA-infection and diabetes. Fourteen (58.3%) of the 24 patients had at least one significant infect exacerbation. In our population, according to the Liou-score (Am J Epidemiol, 2001), estimated 5-survival is high: median 91% (33–100%).
Conclusion: In our adult CF program only 60% of the patients are chronically infected with PA. Most of the patients have a moderate to severe impairment of pulmonary function with at least one significant infect exacerbation per year. Nevertheless, estimated 5-year survival according to the Liou-score is high.